UREA CYCLE DISORDERS (UCD)
FREQUENTLY ASKED QUESTIONS

F.A.Q. About Nutrition Management for Individuals with UCDs[1,2,3] April 2026

NUTRIENT INTAKE	
Can a baby with UCD be breastfed?	Infants with mild or severe UCD may receive breast milk if available, with close monitoring by the metabolic team. For infants with severe UCD, expressed breast milk may be preferred so intake can be measured more precisely. Your team may also recommend supplemental standard infant formula and/or protein-free formula to help meet protein and energy goals consistently. (Rec 2.3)
How much protein can I eat?	Your protein prescription should be individualized based on your age, disease severity, growth, and clinical status. The goal is to provide the highest amount of protein you can safely tolerate while maintaining metabolic stability. Some people with mild UCD may tolerate a protein-controlled diet or even an unrestricted protein diet, while those with more severe UCD often need more restriction. (Rec 2.1)
Do I need medical food (formula)?	Some individuals with UCD, especially those with more severe disease, need essential amino acid-based medical food to help meet protein needs while limiting nitrogen load. Some individuals may also need protein-free medical food to meet energy needs. Your metabolic team will determine whether medical food is needed and how much is appropriate for you.
Why are calories important in UCD?	Adequate calories are important because they help prevent catabolism (muscle breakdown), support growth, and reduce the risk of high ammonia levels. Your metabolic team may recommend higher non-protein calorie intake during times of illness, poor intake, or other metabolic stress.
What if eating is difficult?	For individuals with severe UCD who have persistent poor oral intake, or who need reliable delivery of feeds or medications, tube feeding may be recommended. A feeding specialist may also be involved to support feeding skills and oral development. (Rec 2.4)

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This document is not meant to substitute for the medical advice provided by your doctor.

[1] For children, adolescents, and adults with UCDs, or their caregivers

[2] Based on the 2026 Nutrition Management Guidelines for Urea Cycle Disorders (UCD) by GMDI / MNT4P: https://managementguidelines.net/guidelines_ucd.php

[3] The Management Guidelines Advisory Committee used the nationally standardized condition abbreviation of UCD; curated by the US National Library of Medicine for this and related *guideline products:* https://lhncbc.nlm.nih.gov/

SUPPLEMENTS AND MEDICATIONS	
Do I need vitamin, mineral, or essential fatty acid supplements?	Your metabolic team should regularly assess your intake of vitamins, minerals, and essential fatty acids. Supplements may be recommended based on your diet, medical food use, and blood test results. Common nutrients that may need attention include calcium, copper, iron, selenium, vitamins B12,D, and zinc and essential fatty acids. (Rec 2.5)
Do I need medicines or amino acid supplements to help lower ammonia?	Depending on the type of UCD, your treatment may include nitrogen scavengers and/or amino acid supplements such as L-arginine or L-citrulline. These therapies are used along with nutrition management to help reduce ammonia and support metabolic stability.
MONITORING	
What does my clinic team monitor?	Your clinic team should regularly assess your diet, growth, physical activity level, feeding skills, and nutrition-related physical findings. Monitoring usually also includes blood tests such as plasma amino acids and other labs as needed to assess and guide adjustments to your nutrition plan. (Rec 3.1)
How often will blood tests and clinic visits be needed?	This depends on your age, disease severity, metabolic stability, and life stage. Monitoring is generally more frequent in infancy, in severe UCD, during pregnancy and postpartum, and when metabolic control is not stable. When well, follow-up may range from every few months to yearly, depending on the situation. See Table #2.
What if eating is difficult?	For individuals with severe UCD who have persistent poor oral intake, or who need reliable delivery of feeds or medications, tube feeding may be recommended. A feeding specialist may also be involved to support feeding skills and oral development. (Rec 2.4)

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ILLNESS AND FASTING	
What should I do if I become ill?	You should contact the metabolic clinic when illness occurs and promptly seek medical care if you cannot tolerate feeds or fluids. (Rec 1.7)
What should be in a sick-day plan?	Work with your metabolic team to obtain a written sick-day plan. A sick-day plan should include what to do with diet, fluids, and medications during illness, as well as information on when to call the on-call metabolic specialist and when to seek emergency care. During mild illness, your plan may include temporarily reducing protein intake, increasing calories from non-protein sources, increasing fluids, and continuing prescribed amino acid supplements and nitrogen scavengers as directed. Families should also have an emergency letter or card for urgent medical visits. (Rec 1.7, 1.8)
What if I need surgery, anesthesia, or a prolonged fast?	If you need a medical procedure or any prolonged period without food, it is important to coordinate with your metabolic team in advance. Fasting should be minimized as much as possible, and intravenous (IV) dextrose may be needed before and during the procedure to help prevent catabolism and high ammonia levels. (Rec 2.6)
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WOMEN'S HEALTH, PREGNANCY, AND POSTPARTUM	
Can the menstrual cycle affect UCD symptoms?	Some pubertal, adolescent, and adult females with UCD may have symptoms of metabolic instability that worsen around the menstrual cycle. In those situations, the metabolic team may recommend temporarily increasing energy intake and/or decreasing protein intake. Hormonal birth control may also be considered with the care team.
What should I know about pregnancy?	Women with UCD should work closely with their metabolic team and a high-risk obstetrician before and during pregnancy. Nutrition should be individualized to meet pregnancy needs and prevent catabolism. Nausea and vomiting should be treated aggressively because they can increase metabolic risk. (Rec 7.2, 7.3)
What about labor, delivery, and the postpartum period?	Women with UCD should have a written plan for labor and delivery. Close monitoring is especially important during delivery and the first weeks after birth because the postpartum period carries a high risk for metabolic instability. Energy intake should remain adequate to prevent catabolism. (Rec 7.4, 7.5)
Can women with UCD breastfeed?	Yes. Women with mild or severe UCD who wish to breastfeed should be supported, with close monitoring of nutrient intake and biochemical markers to ensure adequate protein, energy, and fluid intake while maintaining metabolic stability. (Rec 7.5)
LIVER TRANSPLANTATION	
Can liver transplantation be an option for UCD?	For eligible individuals, liver transplantation is an effective treatment option that can improve survival and allow an unrestricted protein diet. Before transplant, the team should complete a nutrition assessment. Around the time of surgery, fasting should be minimized and metabolic support should be carefully managed. After transplant, continued nutrition follow-up is important during recovery and diet transition. (Rec 6.1, 6.2, 6.3)

End of UCD FAQ

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